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## Adults with intellectual disabilities diagnosed with cancer: findings from reviews of their deaths and linked data from the national cancer registry

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# Adults with intellectual disabilities diagnosed with cancer: findings from reviews of their deaths and linked data from the national cancer registry

## TITLE PAGE

### Title

Adults with intellectual disabilities diagnosed with cancer: findings from reviews of their deaths and linked data from the national cancer registry

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The corresponding author attests that all listed authors meet authorship criteria and that no others meeting the criteria have been omitted.

## ABSTRACT

**Objective.** To improve our understanding of cancer in adults with intellectual disabilities, specifically the types of cancer, how cancer was diagnosed, the stage of cancer at diagnosis and the cause of death in people known to have had cancer.

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**Design.** Population-based study using linked data from the Learning Disabilities Mortality Review (LeDeR) programme and the national cancer registry.

**Setting.** England.

**Participants.** 1,096 adults with intellectual disabilities identified by the LeDeR programme who died between 1st January 2017 and 31st December 2019. Linked data from the national cancer registry were available for 771 (70%).

**Outcome measure.** Any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Parts I or II of the Medical Certificate of Cause of Death.

**Results.** More than a third (35%) of people with intellectual disabilities had cancer diagnosed via emergency presentations. Almost half (46%) of cancers were at Stage IV when diagnosed. 36% of deaths were of cancers of the digestive system, almost half of these (48%) were cancer of the colon, rectum or anus. Of those who died with colorectal cancer, 43% were below the age threshold for colorectal screening.

**Conclusions.** This study found differences in the diagnosis of cancer between people with intellectual disabilities and published national data. Symptoms suggestive of cancer were picked up and acted upon in general practice less frequently for people with intellectual disabilities and they were more likely to be diagnosed at a late stage than comparative national data suggests. There is a need for greater awareness of symptoms of cancer in this population, a lower threshold for referral by GPs, and accelerated access to diagnosis and treatment.

**STRENGTHS AND LIMITATIONS OF THE STUDY**

- This study is the first to link data about people with intellectual disabilities known to have had cancer prior to their death with that from the national cancer registry and official cause of death coding from NHS Digital.
- There is an indication that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an ‘unknown’ tumour type.
- The study population was unique in that all died between 2017-2019.
- The small number of deaths in some sub-categories, and when compared with general population data that utilise different baselines, means that conclusions based on these data should be considered tentative.

**MAIN TEXT**

**INTRODUCTION**

We have little contemporary data about the experiences of people with intellectual disabilities who have been diagnosed with cancer.[1] In general, mortality studies of people

with intellectual disabilities indicate a lower proportion of cancer-related deaths than in the general population,[2,3] possibly due to the lower life expectancy of people with intellectual disabilities,[4,5] and cancer being predominantly a disease of older age. Nevertheless, cancer is one of the five most frequently recorded causes of death in studies of adults, or adults and children, with intellectual disabilities,[2,6] and is therefore deserving of attention in any considerations about reducing premature mortality in this population.

Within the population of people with intellectual disabilities, the risk of dying from cancer is not uniform. Females with intellectual disabilities may have increased risk of death from cancer compared to males,[2,7] although this mainly appears to be a feature of younger age groups.[6] There are conflicting findings as to whether overall cancer incidence varies by level of intellectual disability. Landes et al.,[2] reported rates of death from cancer to be higher among adults with mild or moderate intellectual disabilities than in adults with severe or profound and multiple intellectual disability, or those with unspecified intellectual disability. Patja et al.,[8] found there to be no difference in overall cancer incidence by level of intellectual disability.

Conclusions have been drawn about the need to improve preventative measures such as cancer screening [9,10] in people with intellectual disabilities, but there has been little published evidence about the diagnosis, treatment and quality of care provided for people with intellectual disabilities with cancer. In a small study of 11 women with intellectual disabilities who had breast cancer, Satgé et al.,[11] observed that cancers were found at a more advanced stage than in the general population. More recently, a population-based cohort study in the Netherlands concluded that cancer may be underdiagnosed and/or undertreated in people with intellectual disability.[12] A scoping review of cancer treatment and survivorship among people with intellectual and developmental disabilities in the U.S. reported that urgent action was needed to improve collaboration among health care providers.[13]

This paper aims to improve our understanding of cancer in adults with intellectual disabilities who have died and whose deaths were reported to the English Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme. It addresses three research questions:

1. What are the types of cancer diagnosed in adults with intellectual disabilities?
2. How, and at what stage, is cancer diagnosed in adults with intellectual disabilities?
3. What is the cause of death in adults with intellectual disabilities known to have had cancer?

## METHODS

### Study design and data

Data about adults with intellectual disabilities were extracted from LeDeR programme reviews of deaths. The national Learning from Deaths policy[14] requires that all deaths of people with intellectual disabilities (aged 4 years and older) in England should be reported

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to the LeDeR programme and reviewed using LeDeR programme methodology. The review process is described by [Anonymised] et al.[15]

The 10th edition of the International Classification of Diseases (ICD-10) was used to classify diagnoses and causes of death. This is divided into 21 chapters; Chapter 2 relates to neoplasms (cancer) with the codes C00-D49. Data about the ICD-10 codes for causes of death of people with intellectual disabilities notified to the LeDeR programme were obtained from NHS Digital.

For people with intellectual disabilities for whom a LeDeR review of their death had been completed and who were known to have had cancer, data was linked to that held in the national cancer registry in England, which holds information about cancer and tumour diseases. Data linkage was conducted by the National Cancer Registration and Analysis Service (NCRAS) and based on the key identifiers of NHS number and date of birth. Those included were people with intellectual disabilities who died in 2017, 2018 or 2019, for whom a LeDeR review had been completed, and for whom the ICD-10 codes for cause of death had been supplied by NHS Digital. All had cancer listed as a long-term health condition by the reviewer of their death or had cancer included in either Part I or Part II of the Medical Certificate of Cause of Death (MCCD).

The ‘stages’ of cancer, which describe the size of the cancer and its spread, are as follows:

- Stage I – the cancer is small and has not spread.
- Stage II – the cancer has grown but has not spread.
- Stage III – the cancer is larger and may have spread to the surrounding tissues and/or the lymph nodes.
- Stage IV – the cancer has spread from where it started to at least one other body organ; known as "secondary" or "metastatic" cancer.

We used the World Health Organisation definition of the underlying cause of death: the disease or injury which initiated the train of events leading directly to death[16] and the European harmonised definition of avoidable mortality and list of causes of death that has been adopted by the UK Office for National Statistics:[17]

- Preventable mortality. Causes of death that can be mainly avoided through effective public health and primary prevention interventions (i.e., before the onset of diseases/injuries, to reduce incidence).
- Treatable mortality. Causes of death that can be mainly avoided through timely and effective health care interventions, including secondary prevention and treatment (i.e., after the onset of diseases, to reduce case-fatality).
- Avoidable mortality. Avoidable causes of deaths are all those defined as preventable or treatable.

**Outcome**

The outcome of interest was any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Part I or Part II of the MCCD, of deaths occurring during the period 1<sup>st</sup> January 2017 to 31<sup>st</sup> December 2019.

### Exposure

The definition of intellectual disabilities used was the presence of a significantly reduced ability to understand new or complex information, to learn new skills (impaired intelligence), with a reduced ability to cope independently (impaired social functioning) which started before adulthood, with a lasting effect on development.[18]

### Covariates

The covariates and their sources were:

LeDeR review of death: demographic information - age; gender; ethnicity; level of intellectual disabilities; geographic area of residence; usual type of accommodation.

NHS Digital: ICD-10 codes for causes of death recorded on the MCCD.

National cancer registry: relevant timescales (e.g., patient age at diagnosis); how the cancer was diagnosed; the site and stage of the cancer when it was diagnosed.

### Approval for the study

The LeDeR programme had Section 251 approval from the national Health Research Authority's Confidentiality Advisory Group (CAG), on behalf of the Secretary of State, allowing the programme team to handle identifiable data without consent in order to conduct a review of a death, and to link it to NHS Digital cause of death data. An amendment to the agreement (CAG Ref: 20CAG067) enabled the LeDeR programme to link identifiers with national cancer registry data.

The study was funded by NHS England. NHS England had no part in the analysis or presentation of the data.

### Data analysis

Data analysis was undertaken by analysts at [Anonymised]. The analysts worked with the LeDeR team at [Anonymised] to ensure a full understanding of the data and to agree how it was reported. Statistical analyses were carried out using the R programming language. Initial chi-square analyses were carried out on frequency tables as a whole. If there was a significant effect, pairwise chi-square analyses were performed to determine which particular variables had significantly different proportions. For brevity we present only significant pair-wise analyses results.

All numbers fewer than 10 have been suppressed to protect confidentiality.

Where appropriate, data have been compared with general population data, although the difference in baseline numbers mean that comparisons should be interpreted cautiously.

Family members of people with intellectual disabilities were involved in the conceptualisation of the study and with discussing the findings.



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RESULTS

People with intellectual disabilities for whom linked data was not available

Of the 1,096 adults with intellectual disabilities identified by the LeDeR programme, and who met the criteria for inclusion in this study, linked data were available from the national cancer registry for 771 (70%). Thus 325 people were known by the LeDeR programme to have had cancer, but data was not available about them in the national cancer registry. The majority of those for whom data were not available (70%, n=229) had died in 2019, a year for which registrations in the national cancer registry had not yet been completed at the time of writing. Thus, it is likely that these deaths were late registrations rather than omissions from the registry.

For 96 people who died in 2017 or 2018, data were not available from the national cancer registry. These people tended to be older (16% aged 80 or over, compared to 6% of those in the registry); and more had severe or profound and multiple intellectual disabilities (36%) than those in the registry (19%). A quarter (25%) of those not in the cancer registry had an ‘unknown’ tumour type, compared to none of those included.

People with intellectual disabilities for whom linked data were available

Of the 771 people for whom linked data were available, 631 (82%) had cancer reported in Part I of the MCCD; 44 (6%) had cancer recorded as a contributory cause in Part II of the MCCD; and 96 (12%) did not have cancer recorded on the MCCD but had cancer recorded as a long-term condition by the reviewer of their death.

Demographic data for people with cancer (Table 1) were similar as to the population of people with intellectual disabilities whose deaths were reported to the LeDeR programme.[19] The deaths of people with cancer were spread across the geographical regions of England. The greatest proportion was from the Midlands (19%); the least was from the South-west (10%). Males accounted for 54%; females 46%. The majority (80%) of deaths occurred between the ages of 50 and 79 years with small proportions in the youngest and oldest age groups. The median age at death for males was 64 years (SD=13.17) and for females was 62 years (SD=13.54). Almost all (96%) were white British. Almost half (47%) of the 714 people for whom information about level of intellectual disabilities was available had mild intellectual disabilities. Of the 499 people for whom information was available about their usual type of accommodation, more than 80% lived in either supported accommodation (32%), their own or their family home (27%) or a residential home (24%).

Table 1: Demographic information about the people with intellectual disabilities for whom linked data was available

Age Group	LeDeR Data					
	Male		Female		Total†	
	No.	%	No.	%	No.	%
20-34	18	4%	13	4%	31	4%
35-49	29	7%	46	13%	75	10%
50-64	167	40%	154	44%	322	42%

65-79	176	42%	120	34%	296	38%
80+	26	6%	21	6%	47	6%
<i>Total</i>	<i>416</i>	<i>100%</i>	<i>354</i>	<i>100%</i>	<i>771</i>	<i>100%</i>
<i>Unknown/Missing</i>					§	
<b>Median age of death</b>	64		62		63	
<b>Standard Deviation</b>	13.17		13.54		13.34	
<b>Ethnicity</b>						
White British	394	97%	334	95%	728	96%
Non-white British	12	3%	19	5%	32	4%
<i>Total</i>	<i>406</i>	<i>100%</i>	<i>353</i>	<i>100%</i>	<i>759</i>	<i>100%</i>
<i>Unknown/Missing</i>					11	
<b>Intellectual Disability Level</b>						
Mild	181	47%	157	47%	339	47%
Moderate	130	34%	108	33%	238	33%
Severe	58	15%	54	16%	112	16%
Profound / multiple	14	4%	12	4%	26	4%
<i>Total</i>	<i>383</i>	<i>100%</i>	<i>331</i>	<i>100%</i>	<i>715</i>	<i>100%</i>
<i>Unknown/Missing</i>					56	
<b>Geographic Area</b>						
Midlands	82	20%	64	18%	147	19%
North East and Yorkshire	79	19%	44	12%	123	16%
South East	59	14%	64	18%	123	16%
North West	51	12%	60	17%	111	14%
East of England	55	13%	37	10%	92	12%
London	47	11%	49	14%	96	12%
South West	42	10%	37	10%	79	10%
<i>Total</i>	<i>415</i>	<i>100%</i>	<i>355</i>	<i>100%</i>	<i>771</i>	<i>100%</i>
<b>Accommodation</b>						
Supported living	89	32%	73	33%	163	32%
Own or Family	77	28%	60	27%	137	27%
Residential home	70	25%	52	23%	122	24%
Nursing home	34	12%	33	15%	67	13%
Other	§	§	§	§	11	2%
<i>Total</i>	<i>277</i>	<i>100%</i>	<i>222</i>	<i>100%</i>	<i>500</i>	<i>100%</i>
<i>Unknown/Missing</i>					271	

†There is one person whose gender was not recorded. They have been included in the Total column.

§Numbers fewer than 10 have been suppressed.

The types of cancer diagnosed in adults with intellectual disabilities who died from any cause

Information about the type of cancer diagnosed was available for all 771 of those with linked data. Most (89%, n=690) had been diagnosed with one type of cancer; 9% (n=69) with two; 2% (n=12) with three or more different types. Thus, in the 771 people, 865 cancers had been diagnosed.

In males, the most frequently recorded types of cancers were of the digestive organs (28%); skin (12%); lip, oral cavity and pharynx (11%); and male genital organs (10%). In females, the most frequently recorded types of cancer were of the breast (26%); digestive organs (23%); and female genital organs (14%) (Table 2).

Table 2: The most frequently reported cancers by ICD-10-chapter sections, in adults with intellectual disabilities known to have had cancer							
ICD-10 section	Section heading	Males		Females		Total <sup>†</sup>	
		No	%	No	%	No	%
C15-C26	Malignant neoplasms of digestive organs	131	28%	92	23%	224	26%
C50	Malignant neoplasms of breast	§	§	103	26%	105	12%
C43-C44	Melanoma and other malignant neoplasms of skin	56	12%	32	8%	88	10%
C00-C14	Malignant neoplasms of lip, oral cavity and pharynx	52	11%	13	3%	65	8%
C30-C39	Malignant neoplasms of respiratory and intrathoracic organs	39	8%	21	5%	60	7%
C42	Malignant neoplasms of the haemato-poietic and reticuloendothelial system	33	7%	22	6%	55	6%
C51-C58	Malignant neoplasms of female genital organs			54	14%	54	6%
C60-C63	Malignant neoplasms of male genital organs	47	10%			47	5%
C64-C68	Malignant neoplasms of urinary tract	33	7%	13	3%	46	5%
C76-C80	Malignant neoplasms of ill-defined, other secondary and unspecified sites	24	5%	13	3%	37	4%
C81-C96	Malignant neoplasms of lymphoid, hematopoietic and related tissue	19	4%	15	4%	34	4%
C69-C72	Malignant neoplasms of eye, brain and other parts of central nervous system	20	4%	10	3%	30	3%
C40-41; C45-49; C73-75; D00-09; D10-36 D37-48; D49	All other cancers.	11	2%	§	§	20	2%
Total number of cancers		467	100%	397	100%	865	100%
<sup>†</sup> There is one person whose gender was not recorded. They have been included in the Total column.							

<sup>§</sup>Numbers fewer than 10 have been suppressed.

### Route to diagnosis of cancer

Of the 771 people who had been diagnosed with cancer and for whom linked data was available, information about the route to diagnosis was available for 60% (n=462). Cancers in people with intellectual disabilities were more likely to be diagnosed via emergency presentations than general population data suggests: 35% of people with intellectual disabilities had their cancer diagnosed via an emergency referral or attendance, compared to 20% of the general population (Table 3).

**Table 3: Route to diagnosis and stage at diagnosis for people with intellectual disabilities and general population of England**

Route to diagnosis	LeDeR data (2017-2019)		General population data (2016) <sup>‡</sup>	
	No.	%	No.	%
Emergency presentation	162	35%	57,593	20%
GP non-urgent referral	123	27%	72,749	25%
Urgent referral ('two-week wait')	116	25%	115,186	39%
Other (Outpatient attendance/ elective inpatient/ death certificate only)	45	10%	33,343	11%
Screening	16	3%	15,156	5%
<i>Total</i>	462	100%	294,026	100%
<i>Unknown/Missing</i>	309		9,094	
Stage of cancer at diagnosis	LeDeR data (2017-2019)		General population data (2019) <sup>‡</sup>	
	No.	%	No.	%
Stage I	88	18%	90,476	35%
Stage II	78	16%	51,309	20%
Stage III	105	21%	52,399	20%
Stage IV	228	46%	65,803	25%
<i>Total number of cancers</i>	503	100%	259,987	100%
<i>Unknown/Missing/unstageable</i>	362		60,209	
<sup>‡</sup> Data extracted from: <a href="http://www.ncin.org.uk/publications/routes_to_diagnosis">http://www.ncin.org.uk/publications/routes_to_diagnosis</a>				
<sup>‡</sup> Data extracted from: <a href="https://www.cancerdata.nhs.uk/stage_at_diagnosis">https://www.cancerdata.nhs.uk/stage_at_diagnosis</a>				

People with intellectual disabilities diagnosed via an emergency route were disproportionately male (20%, compared to 15% females;  $\chi^2$  (5,  $N=462$ ) = 29.95,  $p=.0042$ ); younger in age (51% in the 20-49 age group, compared to 33% of those in older age groups;  $\chi^2$  (20,  $N=462$ ) = 26.65,  $p=.0017$ ); and living in their own or the family home (36%, compared to 16% of those living in residential care setting;  $\chi^2$  (25,  $N=462$ ) = 64.68,  $p=.0014$ ).

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A significantly smaller proportion of people with intellectual disabilities (25%) had their cancer diagnosed via an urgent general practitioner (GP) referral, (sometimes referred to as a ‘two-week wait’) than did people in the general population (39%).

Sixteen people with intellectual disabilities had their cancer identified by screening. Of these, 14 were diagnosed with breast cancer, a rate comparable with the proportion of cancers identified by screening in the general population.

The stage of cancer when it was diagnosed

Of the 865 cancers diagnosed in people with intellectual disabilities, information about the stage of the cancer when it was diagnosed was available for 58% (n=503). In people with intellectual disabilities, almost half (46%) of cancers diagnosed were at Stage IV, when the cancer had already metastasised, compared to a quarter (25%) of cancers in the general population (Table 3). There was some variation in this in people with intellectual disabilities. Those cancers diagnosed at Stage IV were disproportionately in males (51%, compared to 39% in females;  $X^2(8, N=503) = 15.92, p = .0033$ ); in people in younger age groups (52% aged 20-49 years, compared to 45% in older age groups  $X^2(16, N=503) = 27.18, p = .002$ ); and of cancer of the digestive organs (57% of cancers of the digestive system were diagnosed at Stage IV;  $X^2(56, N=503) = 265.9, p = .0007$ ).

The deaths of adults with intellectual disabilities known to have had cancer

*ICD-10 chapters of underlying causes of death*

Over three-quarters (n=852; 78%) of those known to have had cancer (irrespective of whether linked data from the national cancer registry were available for them), had cancer recorded as their underlying cause of death. Small proportions had respiratory disorders (7%; n=73), circulatory disorders (4%; n=42) or other disorders (11%; n= 124) recorded as their underlying cause of death.

*ICD-10 sections of cancer-related underlying causes of death*

The most frequently recorded ICD-10 section for the cancer-related underlying cause of deaths in people with intellectual disabilities was cancer of the digestive organs (C15-26), reported for over a third (36%) of all deaths from cancer (Table 4). The corresponding proportion in the general population of England was 29%. The second most frequently recorded in people with intellectual disabilities was of ill-defined, secondary and unspecified sites (10%), more than the corresponding proportion in the general population of England (6%), and possibly reflecting the greater than expected proportion of cancers in people with intellectual disabilities that were diagnosed at emergency presentation and in the later stages. The third most frequently recorded in people with intellectual disabilities was of cancer of the respiratory and intrathoracic organs (8%), although this was substantially less than in the general population of England (21%).

There was some variation in the underlying cause of death within the group of people with intellectual disabilities. Cancers of the digestive system were more frequently reported in

males than females (42% males; 30% females;  $X^2(34, N=852) = 209.63, p=.0002$ ); and cancers of the male genital organs were not reported in any males from minority ethnic groups ( $X^2(34, N=852) = 60.262, p=.0011$ ).

We compared the most frequently recorded ICD-10 sections for the cancer-related underlying causes of death for people with intellectual disabilities with those of the general population of England using a chi-square test. There was a significant difference between the population of people with intellectual disabilities and the general population ( $X^2(10, N=852) = 21.79, p=.016$ ). Pairwise testing suggested that in males with intellectual disabilities, genital cancer and respiratory cancer were significantly lower than in males in the general population. In females with intellectual disabilities, respiratory cancer was significantly lower than in females in the general population.

### *Specific ICD-10 codes for cancers of the digestive system*

Disaggregating most of the cancer sections in people with intellectual disabilities was limited by the small numbers in some sections. We therefore focused on cancers of the digestive system, the most frequently reported cancer-related underlying cause of death in people with intellectual disabilities and more frequently reported in people with intellectual disabilities than the general population. Three types of cancer accounted for 82% of cancers of the digestive system in people with intellectual disabilities (Table 5). The most frequently recorded was cancer of the colon, rectum and anus – almost half (48%) of all cancers of the digestive system were in these sites, a significantly greater proportion than the 34% in the general population ( $X^2(5, N=309) = 21.52, p=.00064$ ). The second most frequently recorded was cancer of the oesophagus (19% of all cancers of the digestive system), slightly more but not significantly so to the 16% in the general population. Third was cancer of the pancreas (15% of all cancers of the digestive system), slightly less, but not significantly so, than the proportion in the general population (20%).

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Table 4: The most frequently recorded ICD-10 codes for the cancer-related underlying causes of death for people with intellectual disabilities and general population of England													
		LeDeR data (2017-2019) (age 18+)						General population (2018) (age 20+) <sup>±</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
ICD-10 codes	Neoplasm of...	no.	%	no.	%	no.	%	no.	%	no.	%	no.	%
C15-C26	Digestive organs	191	42%	117	30%	309	36%	23,297	32%	15,953	26%	39,250	29%
C76-C80	Ill-defined, other secondary & unspecified sites	53	12%	32	8%	85	10%	4,003	6%	4,448	7%	8,451	6%
C30-C39	Respiratory & intrathoracic organs	46	10%	23	6%	69	8%	15,708	22%	12,910	21%	28,618	21%
C50	Breast	§	§	67	17%	69	8%	78	<1%	9,562	15%	9,640	7%
C81-C96; C42	Lymphoid, hematopoietic & related tissue	37	8%	27	7%	64	8%	3,799	5%	3,233	5%	7,032	5%
C51-C58	Female genital organs			60	15%	60	7%			6,818	11%	6,818	5%
C64-C68	Urinary tract	32	7%	18	5%	50	6%	5,492	8%	2,844	5%	8,336	6%
C69-C72	Eye, brain & other central nervous system	24	5%	12	3%	36	4%	2,200	3%	1,603	3%	3,803	3%
C60-C63	Male genital organs	34	7%			34	4%	10,243	14%			10,243	8%
C43-C44	Melanoma & other skin	17	4%	13	3%	30	4%	1671	2%	1,007	2%	2,678	2%
C00-14; C40-41; C45-49; C73-75; D00-09;	All other causes	22	5%	24	6%	46	5%	5,935	8%	3,691	5%	9,626	8%

D10-36; D37-48.													
<b>Total</b>		458	100%	393	100%	852	100%	72,426	100%	62,069	100%	134,495	100%
<sup>†</sup> There is one person whose gender was not recorded. They have been included in the Total column. <sup>‡</sup> Data extracted from <a href="https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018">https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018</a> <sup>§</sup> Numbers fewer than 10 have been suppressed.													



**Table 5: The most frequently recorded ICD-10 codes for underlying cause of death of cancer of the digestive system for people with intellectual disabilities and general population of England**

		LeDeR data (age 18+)						England (2018) (age 20+) <sup>‡</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
ICD-10 codes	Neoplasm of...	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers
C18-C21	Colon, rectum and anus	86	45%	60	51%	147	48%	7,509	32%	6,387	37%	13,896	34%
C15	Oesophagus	43	23%	16	14%	59	19%	4,564	20%	1,985	11%	6,549	16%
C25	Pancreas	27	14%	18	15%	45	15%	4,163	18%	3,868	22%	8,031	20%
C22	Liver and intrahepatic bile ducts	15	8%	§	§	23	7%	2,907	12%	1,794	11%	4,701	12%
C16	Stomach	12	6%	§	§	17	6%	2,197	9%	1,213	7%	3,410	8%
C17 C23 C24 C26	All other digestive organs	§	§	10	4%	18	6%	1,957	8%	2,031	12%	3,988	10%
C15-C26	Digestive organs	191	100%	117	100%	309	100%	23,297	100%	17,278	100%	40,575	100%
<sup>†</sup> There is one person whose gender was not recorded. They have been included in the Total column.													
<sup>‡</sup> Data extracted from <a href="https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018">https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018</a>													
§Numbers fewer than 10 have been suppressed.													

### *Potentially avoidable deaths from cancer*

For those who died with cancer as an underlying cause of death, we examined the proportion of cancers known to be avoidable (either preventable or treatable), using the harmonised definition of avoidable mortality and a list of causes of death considered to be avoidable.[17] Overall, 19% (n=163) people with intellectual disabilities had a type of cancer that was a preventable cause of death. The largest proportion was due to cancer of the oesophagus (n=52; 32% of all preventable causes of death from cancer) and the lung (n=44; 27% of all preventable causes of death from cancer). 23% (n=200) of people with intellectual disabilities had a type of cancer that was a treatable underlying cause of death. The largest proportion was due to colorectal cancer (n=106; 53% of all treatable causes of death from cancer) and breast cancer (in females only) (n=57; 29% of all treatable causes of death from cancer).

## **DISCUSSION**

Despite the first major study of cancer in people with intellectual disabilities in England being conducted in 1997, the lack of contemporary data leads to significant policy and practice gaps. This study aimed to extend our knowledge about cancer in people with intellectual disabilities in England. One of the strengths of our study is that it links data about people with intellectual disabilities known to have had cancer prior to their death with data from the national cancer registry and official cause of death coding from NHS Digital. This provides more comprehensive information than would otherwise be available from any of the data sources alone. There is an indication, however, that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an 'unknown' tumour type. The study population was unique in that all died between 2017-2019; it was not a cross-sectional cohort of the population, so comparison with other findings need to be made with this caveat in mind. In addition, the small number of deaths in some sub-categories, and when compared with general population data that utilise different baselines, means that conclusions based on these data should be considered tentative.

Overall, people with intellectual disabilities more frequently had cancer diagnosed via emergency presentations and less frequently via primary care than did people in the general population, suggesting that symptoms suggestive of cancer are picked up and acted upon in general practice less frequently for people with intellectual disabilities. Almost half (46%) of cancers in people with intellectual disabilities were diagnosed at Stage IV when the cancer had already metastasised to other parts of the body. Cancer survival data emphasise the importance of early diagnosis and that for most cancers, survival at one and five years is much higher if the cancer is detected at Stage I than if it is detected later.[20] The NHS Long Term Plan[21] states the ambition that by 2028, the proportion of cancers diagnosed at Stages I and II will rise to three-quarters of cancer patients. With only a third (34%) of cancers in people with intellectual disabilities identified at these stages, there is clearly

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much work to do to raise greater awareness of symptoms of cancer in this population, lower the threshold for referral by GPs, and accelerate access to diagnosis and treatment.

Digestive system cancers were the most common type of cancer death in this study, echoing the findings of other studies.[6,8,22] This may be influenced by gastrointestinal tract dysfunction, gastro-oesophageal reflux disease, and chronic constipation which are common in people with intellectual disabilities.[23] There is also a suggestion that a higher incidence of these cancers in people with intellectual disabilities, compared to the general population, may be associated with genetic deletions and family history, being overweight, inactive, or having poor nutrition.[24] A greater awareness of this amongst support staff and health professionals is important so that preventative measures can be instigated such as caregivers being vigilant about bowel habits and implementing interventions to increase fibre intake and exercise in daily activities.[24]

Colorectal screening presents an opportunity to discover early colorectal cancer and is available to everyone over the age of 60 years in England, with the programme expanding to include 56-year-olds from 2021. Although screening rates for colorectal cancer in people with intellectual disabilities are approaching those of the general population[25] our study found that 43% of people diagnosed with colorectal cancer were aged 18-59 years, suggesting that the age threshold for colorectal screening in people with intellectual disabilities may need reducing.

After colorectal cancer, breast cancer was the second most frequently reported treatable cause of death in our population. Although rates of breast cancer appear to be similar in women with and without intellectual disabilities, under-utilisation of breast cancer screening in people with intellectual disabilities[25] may reduce the number identified. Nulliparity, being overweight, and a lack of exercise are known factors that increase breast cancer risk[26] and are particularly pertinent to people with intellectual disabilities. Research is sparse about the treatment options and decision-making process for women with intellectual disabilities who have breast cancer; research relating to disabled women in general suggests that they are less likely to undergo breast-conserving surgery and are less likely to receive neoadjuvant chemotherapy and radiotherapy than their non-disabled peers.[27]

Some of the factors influencing disparities in cancer outcomes will be related to the social and economic context that shapes a person's ability to access cancer care. Mechanisms leading to poorer outcomes in people with intellectual disabilities have been identified as provider bias and ableism;[28] negative attitudes;[29] the invisibility of people with intellectual disabilities;[30] diagnostic overshadowing in which symptoms are misinterpreted as due to a person's behaviour, mental state or communication impairments;[31] and unequal access to health services.[32] Preventative measures to reduce the risk of cancer, screening and health checks to identify it early, transparent decision-making processes about options available, and access to diagnostic and treatment interventions that provide a chance of optimal outcomes for people with cancer are all

needed. So too, is a closer research gaze on the quality of care provided to people with intellectual disabilities who have cancer in order to ensure their equitable access to services.

## ACKNOWLEDGEMENTS

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## COMPETING INTERESTS

The authors have declared no competing interests.

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For peer review only



STROBE Statement—Checklist of items that should be included in reports of *cross-sectional studies*

	Item No	Recommendation	Page No
Title and abstract	1	(a) Indicate the study’s design with a commonly used term in the title or the abstract	Abstract Line 4 (p.2)
		(b) Provide in the abstract an informative and balanced summary of what was done and what was found	Abstract
Introduction			
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported	p.2-3
Objectives	3	State specific objectives, including any prespecified hypotheses	p.3
Methods			
Study design	4	Present key elements of study design early in the paper	p.3-4
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection	p.3-4
Participants	6	(a) Give the eligibility criteria, and the sources and methods of selection of participants	p.3-4
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable	p.3-4
Data sources/measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group	p.5
Bias	9	Describe any efforts to address potential sources of bias	p.15
Study size	10	Explain how the study size was arrived at	p.4-5
Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why	p.3
Statistical methods	12	(a) Describe all statistical methods, including those used to control for confounding	p.5
		(b) Describe any methods used to examine subgroups and interactions	p.5
		(c) Explain how missing data were addressed	p.5
		(d) If applicable, describe analytical methods taking account of sampling strategy	n/a
		(e) Describe any sensitivity analyses	n/a
Results			
Participants	13*	(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed	p.6-15
		(b) Give reasons for non-participation at each stage	n/a
		(c) Consider use of a flow diagram	n/a
Descriptive data	14*	(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders	p.6-7
		(b) Indicate number of participants with missing data for each variable of interest	p.6-15



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Outcome data	15*	Report numbers of outcome events or summary measures	p.6-15
Main results	16	(a) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included	p.6-15
		(b) Report category boundaries when continuous variables were categorized	n/a
		(c) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period	n/a
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses	p.6-15
<b>Discussion</b>			
Key results	18	Summarise key results with reference to study objectives	p.13-15
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias	p.15-17
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence	p.15-17
Generalisability	21	Discuss the generalisability (external validity) of the study results	p.15-17
<b>Other information</b>			
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based	p.17

\*Give information separately for exposed and unexposed groups.

**Note:** An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at <http://www.plosmedicine.org/>, Annals of Internal Medicine at <http://www.annals.org/>, and Epidemiology at <http://www.epidem.com/>). Information on the STROBE Initiative is available at [www.strobe-statement.org](http://www.strobe-statement.org).

# BMJ Open

## Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

Journal:	<i>BMJ Open</i>
Manuscript ID	bmjopen-2021-056974.R1
Article Type:	Original research
Date Submitted by the Author:	10-Jan-2022
Complete List of Authors:	Heslop, Pauline; University of Bristol Faculty of Social Sciences and Law, Norah Fry Centre for Disability Studies Cook, Adam; NHS South Central and West Commissioning Support Unit South Regional Office Sullivan, Brian; University of Bristol Faculty of Health Sciences Calkin, Rachel; University of Bristol Faculty of Social Sciences and Law Pollard, Johanna; University of Bristol Faculty of Social Sciences and Law Byrne, Victoria; University of Bristol Faculty of Social Sciences and Law
<b>Primary Subject Heading</b>:	General practice / Family practice
Secondary Subject Heading:	Medical management
Keywords:	Adult oncology < ONCOLOGY, Health policy < HEALTH SERVICES ADMINISTRATION & MANAGEMENT, PRIMARY CARE

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# Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

## TITLE PAGE

### Title

Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

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**ABSTRACT**

**Objective.** To improve our understanding of cancer in adults with intellectual disabilities.

**Design.** Population-based study using linked data about deceased adults from the Learning [Intellectual] Disabilities Mortality Review (LeDeR) programme, the national cancer registry, and NHS Digital.

**Setting.** England.

**Participants.** 1,096 adults with intellectual disabilities identified by the LeDeR programme who died between 1st January 2017 and 31st December 2019.

**Outcome measure.** Any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Parts I or II of the Medical Certificate of Cause of Death.

**Results.** In decedents with intellectual disabilities and cancer, more than a third (35%; n=162) had cancer diagnosed via emergency presentations. Almost half (45%; n=228) of cancers were at Stage IV when diagnosed. More than a third (36%; n=309) of underlying causes of deaths were of cancers of the digestive system; almost half of these (48%; n=147) were cancer of the colon, rectum or anus. Of those who died with colorectal cancer, 43% were below the age threshold for colorectal screening.

**Conclusions.** In decedents with intellectual disabilities, symptoms suggestive of cancer had tended to be identified most frequently as an emergency and at a late stage. There is a need for greater awareness of symptoms of cancer in this population, a lower threshold for referral by GPs, accelerated access to diagnosis and treatment, and consideration paid to lowering the age for colorectal screening.

**STRENGTHS AND LIMITATIONS OF THE STUDY**

- This study is the first to link data about adults with intellectual disabilities known to have had cancer prior to their death with that from the national cancer registry and official cause of death coding from NHS Digital.
- There is an indication that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an ‘unknown’ tumour type.
- There was limited published data available about decedents with cancer, so comparing our data to general population data was not always possible.
- The small number of deaths in some sub-categories, and when compared with general population data about underlying causes of death means that conclusions based on these data should be considered tentative.

## MAIN TEXT

### INTRODUCTION

We have little contemporary data about the experiences of adults with intellectual disabilities who have been diagnosed with cancer.[1] In general, mortality studies of people with intellectual disabilities indicate a lower proportion of cancer-related deaths than in the general population,[2,3] possibly due to the lower life expectancy of people with intellectual disabilities,[4,5] and cancer being predominantly a disease of older age. Nevertheless, cancer is one of the five most frequently recorded causes of death in studies of adults, or adults and children, with intellectual disabilities,[2,6] and is therefore deserving of attention in any considerations about reducing premature mortality in this population.

Within the population of people with intellectual disabilities, the risk of dying from cancer is not uniform. Females with intellectual disabilities may have increased risk of death from cancer compared to males,[2,7] although this mainly appears to be a feature of younger age groups.[6] There are conflicting findings as to whether overall cancer incidence varies by level of intellectual disability. Landes et al.,[2] reported rates of death from cancer to be higher among adults with mild or moderate intellectual disabilities than in adults with severe or profound and multiple intellectual disability, or those with unspecified intellectual disability. Patja et al.,[8] found there to be no difference in overall cancer incidence by level of intellectual disability.

Conclusions have been drawn about the need to improve preventative measures such as cancer screening [9,10] in people with intellectual disabilities, but there has been little published evidence about the diagnosis, treatment and quality of care provided for people with intellectual disabilities with cancer. In a small study of 11 women with intellectual disabilities who had breast cancer, Satgé et al.,[11] observed that cancers were found at a more advanced stage than in the general population. More recently, a population-based cohort study in the Netherlands concluded that cancer may be underdiagnosed and/or undertreated in people with intellectual disability.[12] A scoping review of cancer treatment and survivorship among people with intellectual and developmental disabilities in the U.S. reported that urgent action was needed to improve collaboration among health care providers.[13]

This paper aims to improve our understanding of cancer in adults with intellectual disabilities who have died and whose deaths were reported to the English Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme. It addresses three research questions:

1. What are the types of cancer diagnosed in adults with intellectual disabilities who have died?
2. How, and at what stage, was cancer diagnosed in adults with intellectual disabilities who have died?

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3. What is the underlying cause of death in adults with intellectual disabilities known to have had cancer?

**METHODS**

**Study design and data**

Data about deceased adults with intellectual disabilities were extracted from the LeDeR programme reviews of deaths. The national Learning from Deaths policy[14] requires that all deaths of people with intellectual disabilities (aged 4 years and older) in England should be reported to the LeDeR programme and reviewed using LeDeR programme methodology. The review process is described by [Anonymised] et al.[15]

Data about the ICD-10 codes for causes of death of people with intellectual disabilities notified to the LeDeR programme were obtained from NHS Digital. Data linkage was conducted by NHS Digital and based on the key identifiers of NHS number, date of birth and date of death.

For adults with intellectual disabilities for whom a LeDeR review of their death had been completed and who were known to have had cancer, data was linked to that held in the national cancer registry in England, which holds information about cancer and tumour diseases. Data linkage was conducted by the National Cancer Registration and Analysis Service (NCRAS) and based on the key identifiers of NHS number and date of birth. Those included were adults with intellectual disabilities who died in 2017, 2018 or 2019, for whom a LeDeR review had been completed, and for whom the ICD-10 codes for cause of death had been supplied by NHS Digital. All had cancer listed as a long-term health condition by the reviewer of their death or had cancer included in either Part I or Part II of the Medical Certificate of Cause of Death (MCCD).

Data about decedents in the general population of England were drawn from data already published by the Office for National Statistics in the population data series about deaths, and Public Health England cancer mortality data. Comparative data about other aspects covered in this paper were not available for decedents in England.

The 10th edition of the International Classification of Diseases (ICD-10) was used to classify diagnoses and causes of death. This is divided into 21 chapters; Chapter 2 relates to neoplasms (cancer) with the codes C00-D49.

The ‘stages’ of cancer, which describe the size of the cancer and its spread, are as described by Cancer Research UK[16] as follows:

- Stage I – the cancer is small and has not spread.
- Stage II – the cancer has grown but has not spread.
- Stage III – the cancer is larger and may have spread to the surrounding tissues and/or the lymph nodes.

- Stage IV – the cancer has spread from where it started to at least one other body organ; known as "secondary" or "metastatic" cancer.

Stage 0 refers to 'carcinoma in situ', 'precancerous changes' or 'non-invasive cancer'. Many of these will never develop into cancer and for this reason we have not included them in this study.

We used the World Health Organisation definition of the underlying cause of death: the disease or injury which initiated the train of events leading directly to death[21]. Although we are aware that the underlying cause of death in some people with intellectual disabilities may be inaccurately reported [18, 19, 20], such evidence does not specifically implicate the reporting of deaths from cancer, so we did not amend any cause of death reports.

We used the European harmonised definition of avoidable mortality and list of causes of death that has been adopted by the UK Office for National Statistics:[21]

- Preventable mortality. Causes of death that can be mainly avoided through effective public health and primary prevention interventions (i.e., before the onset of diseases/injuries, to reduce incidence).
- Treatable mortality. Causes of death that can be mainly avoided through timely and effective health care interventions, including secondary prevention and treatment (i.e., after the onset of diseases, to reduce case-fatality).
- Avoidable mortality. Avoidable causes of deaths are all those defined as preventable or treatable.

## Outcome

The outcome of interest was any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Part I or Part II of the MCCD, of deaths occurring during the period 1<sup>st</sup> January 2017 to 31<sup>st</sup> December 2019.

## Exposure

The definition of intellectual disabilities used was the presence of a significantly reduced ability to understand new or complex information, to learn new skills (impaired intelligence), with a reduced ability to cope independently (impaired social functioning) which started before adulthood, with a lasting effect on development.[22]

## Covariates

The covariates and their sources were:

LeDeR review of death: demographic information - age; gender; ethnicity; level of intellectual disabilities; geographic area of residence; usual type of accommodation.

NHS Digital: ICD-10 codes for causes of death recorded on the MCCD.

National cancer registry: relevant timescales (e.g., patient age at diagnosis); how the cancer was diagnosed; the site and stage of the cancer when it was diagnosed.



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**Ethical approval for the study**

The LeDeR programme had Section 251 approval from the national Health Research Authority’s Confidentiality Advisory Group (CAG), on behalf of the Secretary of State, allowing the programme team to handle identifiable data without consent in order to conduct a review of a death, and to link it to NHS Digital cause of death data. An amendment to the agreement (CAG Ref: 20CAG067) enabled the LeDeR programme to link identifiers with national cancer registry data.

The study was funded by NHS England. NHS England had no part in the analysis or presentation of the data.

**Patient and public involvement**

Family members of people with intellectual disabilities were involved in the conceptualisation of the study and in discussing the findings.

**Data analysis**

Data analysis was undertaken by analysts at [Anonymised]. The analysts worked with the LeDeR team at [Anonymised] to ensure a full understanding of the data and to agree how it was reported. Statistical analyses were carried out using the R programming language. Initial chi-square analyses were carried out on frequency tables as a whole. If there was a significant effect, pairwise chi-square analyses were performed to determine which particular variables had significantly different proportions. For brevity we present only significant pair-wise analyses results.

All numbers fewer than 10 have been suppressed to protect confidentiality.

Where appropriate, data about underlying causes of death have been compared with published general population data.

We present the findings for three distinct but inter-related groups of adults with intellectual disabilities:

Group 1: Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1,096).

Group 2: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771).

Group 3: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852).

**RESULTS**

Demographic data about adults with intellectual disabilities included in each group of the study are presented in Table 1. Overall, there was little difference in the demographic information relating to those in Groups 1, 2 and 3, although the median age at death of those in Group 3 was slightly lower than that of Group 1 and Group 2.

**Table 1: Demographic information about those included in the study**

<b>Group 1:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1,096).						
<b>Group 2:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771).						
<b>Group 3:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852).						
	<b>Group 1</b>		<b>Group 2</b>		<b>Group 3</b>	
	<b>No.</b>	<b>%</b>	<b>No.</b>	<b>%</b>	<b>No.</b>	<b>%</b>
<b>Gender†</b>						
Males	581	53%	416	54%	457	54%
Females	514	47%	354	46%	394	46%
<b>Age Group</b>						
20-34	44	4%	31	4%	38	4%
35-49	123	11%	75	10%	104	12%
50-64	430	39%	322	42%	369	43%
65-79	420	38%	296	38%	300	35%
80+	79	7%	47	6%	41	5%
<i>Total</i>	<i>1096</i>	<i>100%</i>	<i>771</i>	<i>100%</i>	<i>852</i>	<i>100%</i>
<b>Median age of death</b>	63		63		61	
<b>Standard Deviation</b>	13.33		13.34		12.78	
<b>Ethnicity</b>						
White British	1034	96%	728	96%	803	96%
Non-white British	45	4%	32	4%	35	4%
<i>Total</i>	<i>1079</i>	<i>100%</i>	<i>760</i>	<i>100%</i>	<i>838</i>	<i>100%</i>
<i>Unknown/Missing</i>	<i>17</i>		<i>11</i>		<i>14</i>	
<b>Level of Intellectual Disability</b>						
Mild	453	44%	339	47%	360	45%
Moderate	345	34%	238	33%	268	34%
Severe	183	18%	112	16%	140	18%
Profound / multiple	44	4%	26	4%	31	4%
<i>Total</i>	<i>1025</i>	<i>100%</i>	<i>715</i>	<i>100%</i>	<i>799</i>	<i>100%</i>
<i>Unknown/Missing</i>	<i>71</i>		<i>56</i>		<i>53</i>	
<b>Geographic Area</b>						
Midlands	214	20%	147	19%	171	20%
North East and Yorkshire	168	15%	123	16%	131	15%
South East	181	17%	123	16%	147	17%
North West	160	15%	111	14%	125	15%
East of England	139	13%	92	12%	107	13%
London	128	12%	96	12%	94	11%
South West	106	10%	79	10%	77	9%
<i>Total</i>	<i>1096</i>	<i>100%</i>	<i>771</i>	<i>100%</i>	<i>852</i>	<i>100%</i>
<b>Accommodation</b>						
Supported living	258	33%	163	32%	198	32%
Own or Family	207	27%	137	27%	175	28%
Residential home	190	24%	122	24%	145	23%
Nursing home	109	14%	67	13%	89	14%
Other	14	2%	11	2%	11	2%
<i>Total</i>	<i>778</i>	<i>100%</i>	<i>500</i>	<i>100%</i>	<i>618</i>	<i>100%</i>

<i>Unknown/Missing</i>	318	271	234
†There is one person whose gender was not recorded. §Numbers fewer than 10 have been suppressed.			

**Adults with intellectual disabilities known from LeDeR data to have died with cancer (Group 1)**

There were 1,096 adults with intellectual disabilities who were known from LeDeR data to have died with cancer. Demographic data for adults with intellectual disabilities in Group 1 were similar to the overall adult population of people with intellectual disabilities whose deaths were reported to the LeDeR programme in 2018.[23]

Most (70%; n=771) of those included in Group 1 had linked data available from the national cancer registry and form Group 2. However, 325 adults were known by the LeDeR programme to have died with cancer, but data was not available about them in the national cancer registry.

The majority of those for whom data in the national cancer registry were not available (70%, n=229) had died in 2019, a year for which registrations in the national cancer registry had not yet been completed at the time of writing. Thus, it is likely that these deaths were late registrations rather than omissions from the registry.

Data were not available from the national cancer registry for 96 adults who died in 2017 or 2018 and who were known by the LeDeR programme to have had cancer. These people tended to be older (16% aged 80 or over, compared to 6% of those in the registry); and more had severe or profound and multiple intellectual disabilities (36%) than those in the registry (19%). A quarter (25%) of those not in the cancer registry had an ‘unknown’ tumour type, compared to none of those included.

**Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2)**

There were 771 adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2).

Information about the type of cancer diagnosed was available for all 771 of those in Group 2 (Table 2). Most (89%, n=690) had been diagnosed with one type of cancer; 9% (n=69) with two; 2% (n=12) with three or more different types. Thus, in the 771 people, 865 cancers had been diagnosed.

In males, the most frequently recorded types of cancers were of the digestive organs (28%); skin (12%); lip, oral cavity and pharynx (11%); and male genital organs (10%). In females, the most frequently recorded types of cancer were of the breast (26%); digestive organs (23%); and female genital organs (14%).

**Table 2: The most frequently reported cancers by ICD-10-chapter sections in adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2) (n=771)**

ICD-10 section	Section heading	Males		Females		Total <sup>†</sup>	
		No	%	No	%	No	%
C15-C26	Malignant neoplasms of digestive organs	131	28%	92	23%	224	26%
C50	Malignant neoplasms of breast	§	§	103	26%	105	12%
C43-C44	Melanoma and other malignant neoplasms of skin	56	12%	32	8%	88	10%
C00-C14	Malignant neoplasms of lip, oral cavity and pharynx	52	11%	13	3%	65	8%
C30-C39	Malignant neoplasms of respiratory and intrathoracic organs	39	8%	21	5%	60	7%
C42	Malignant neoplasms of the haemato-poietic and reticuloendothelial system	33	7%	22	6%	55	6%
C51-C58	Malignant neoplasms of female genital organs			54	14%	54	6%
C60-C63	Malignant neoplasms of male genital organs	47	10%			47	5%
C64-C68	Malignant neoplasms of urinary tract	33	7%	13	3%	46	5%
C76-C80	Malignant neoplasms of ill-defined, other secondary and unspecified sites	24	5%	13	3%	37	4%
C81-C96	Malignant neoplasms of lymphoid, hematopoietic and related tissue	19	4%	15	4%	34	4%
C69-C72	Malignant neoplasms of eye, brain and other parts of central nervous system	20	4%	10	3%	30	3%
C40-41; C45-49; C73-75; D00-09; D10-36 D37-48; D49	All other cancers.	11	2%	§	§	20	2%
<b>Total number of cancers in the 771 people</b>		<b>467</b>	<b>100%</b>	<b>397</b>	<b>100%</b>	<b>865</b>	<b>100%</b>

<sup>†</sup>There is one person whose gender was not recorded. They have been included in the Total column.  
<sup>§</sup>Numbers fewer than 10 have been suppressed.

Of the 771 adults with intellectual disabilities in Group 2, information about the route to diagnosis was available for 60% (n=462). In these 462 adults with intellectual disabilities, cancers were more likely to be diagnosed via emergency presentations than any other route: 35% of adults with intellectual disabilities had their cancer diagnosed via an

emergency referral or attendance, compared to 27% diagnosed via a non-urgent referral by their GP, and 25% diagnosed via an urgent referral (Table 3).

Table 3: Route to diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry		
	LeDeR data (2017-2019)	
	No.	%
Emergency presentation	162	35%
GP non-urgent referral	123	27%
Urgent referral ('two-week wait')	116	25%
Other (Outpatient attendance/ elective inpatient/ death certificate only)	45	10%
Screening	16	3%
Total	462	100%
Unknown/Missing	309	

Adults with intellectual disabilities who were diagnosed via an emergency route were disproportionately male (20%, compared to 15% females;  $\chi^2(5, N=462) = 29.95, p=.0042$ ); younger in age (51% in the 20-49 age group, compared to 33% of those in older age groups;  $\chi^2(20, N=462) = 26.65, p=.0017$ ); and living in their own or the family home (36%, compared to 16% of those living in residential care setting;  $\chi^2(25, N=462) = 64.68, p=.0014$ ).

Sixteen adults with intellectual disabilities had their cancer identified by screening. Of these, 14 were diagnosed with breast cancer, a rate comparable with the proportion of cancers identified by screening in the general population.

Of the 865 cancers diagnosed in the 771 adults with intellectual disabilities in Group 2, information about the stage of the cancer when it was diagnosed was available for 58% (n=502) of the different cancers. Almost half (46%) of the cancers diagnosed were at Stage IV when the cancer had already metastasised (Table 4).

Table 4: Stage of cancer at diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry		
	LeDeR data (2017-2019)	
	No.	%
Stage I	88	18%
Stage II	78	16%
Stage III	105	21%
Stage IV	228	45%

Total number of cancers	502	100%
Unknown/Missing/unstageable	363	

There was some variation in the stage of cancer in adults with intellectual disabilities in Group 2. Those cancers diagnosed at Stage IV were disproportionately in males (51%, compared to 39% in females;  $\chi^2 (8, N=503) = 15.92, p=.0033$ ); in adults in younger age groups (52% aged 20-49 years, compared to 45% in older age groups  $\chi^2 (16, N=503) = 27.18, p=.002$ ); and of cancer of the digestive organs (57% of cancers of the digestive system were diagnosed at Stage IV;  $\chi^2 (56, N=503) = 265.9, p=.0007$ ).

### **Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (Group 3)**

#### *ICD-10 chapters of underlying causes of death*

Over three-quarters ( $n=852$ ; 78%) of the 1,096 adults with intellectual disabilities in Group 1 had cancer recorded as their underlying cause of death in the official cause of death coding received from NHS Digital (small proportions had respiratory disorders (7%;  $n=73$ ), circulatory disorders (4%;  $n=42$ ) or other disorders (11%;  $n=124$ ) recorded as their underlying cause of death).

#### *ICD-10 sections of cancer-related underlying causes of death*

The most frequently recorded ICD-10 section for the cancer-related underlying cause of deaths in the 852 adults in Group 3 was cancer of the digestive organs (C15-26), reported for over a third (36%) of deaths (Table 5). The corresponding proportion in decedents in the general population of England was 29%. The second most frequently recorded in adults with intellectual disabilities in Group 3 was of ill-defined, secondary and unspecified sites (10%), more than the corresponding proportion in the general population of England (6%), and possibly reflecting the greater than expected proportion of cancers in adults with intellectual disabilities that were diagnosed at emergency presentation and in the later stages. The third most frequently recorded in adults with intellectual disabilities in Group 3 was of cancer of the respiratory and intrathoracic organs (8%), although this was substantially less than in the general population of England (21%).

There was some variation within the adults with intellectual disabilities in Group 3 according to the underlying cause of death. Cancers of the digestive system were more frequently reported in males than females (42% males; 30% females;  $\chi^2 (34, N=852) = 209.63, p=.0002$ ); and cancers of the male genital organs were not reported in any males from minority ethnic groups ( $\chi^2 (34, N=852) = 60.262, p=.0011$ ).

We compared the most frequently recorded ICD-10 sections for the cancer-related underlying causes of death for adults with intellectual disabilities in Group 3 with those of the general population of England using a chi-square test. There was a significant difference

Table 5: The most frequently recorded ICD-10 codes for cancer-related underlying causes of death in adults with intellectual disabilities for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (Group 3), compared to data reported for the general population of England													
		LeDeR data (2017-2019) (age 18+)						General population (2018) (age 20+) <sup>±</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
ICD-10 codes	Neoplasm of...	no.	%	no.	%	no.	%	no.	%	no.	%	no.	%
C15-C26	Digestive organs	191	42%	117	30%	309	36%	23,297	32%	15,953	26%	39,250	29%
C76-C80	Ill-defined, other secondary & unspecified sites	53	12%	32	8%	85	10%	4,003	6%	4,448	7%	8,451	6%
C30-C39	Respiratory & intrathoracic organs	46	10%	23	6%	69	8%	15,708	22%	12,910	21%	28,618	21%
C50	Breast	§	§	67	17%	69	8%	78	<1%	9,562	15%	9,640	7%
C81-C96; C42	Lymphoid, hematopoietic & related tissue	37	8%	27	7%	64	8%	3,799	5%	3,233	5%	7,032	5%
C51-C58	Female genital organs			60	15%	60	7%			6,818	11%	6,818	5%
C64-C68	Urinary tract	32	7%	18	5%	50	6%	5,492	8%	2,844	5%	8,336	6%
C69-C72	Eye, brain & other central nervous system	24	5%	12	3%	36	4%	2,200	3%	1,603	3%	3,803	3%
C60-C63	Male genital organs	34	7%			34	4%	10,243	14%			10,243	8%
C43-C44	Melanoma & other skin	17	4%	13	3%	30	4%	1671	2%	1,007	2%	2,678	2%
C00-14; C40-41;	All other causes	22	5%	24	6%	46	5%	5,935	8%	3,691	5%	9,626	8%



C45-49; C73-75; D00-09; D10-36; D37-48.													
<b>Total</b>		458	100%	393	100%	852	100%	72,426	100%	62,069	100%	134,495	100%
<sup>†</sup> There is one person whose gender was not recorded. They have been included in the Total column. <sup>‡</sup> Data extracted from <a href="https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018">https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018</a> <sup>§</sup> Numbers fewer than 10 have been suppressed.													



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between the population of adults with intellectual disabilities and adults in the general population ( $X^2(10, N=852) = 21.79, p=.016$ ). Pairwise testing suggested that in males with intellectual disabilities, genital cancer and respiratory cancer were significantly lower than in males in the general population. In females with intellectual disabilities, respiratory cancer was significantly lower than in females in the general population.

*Specific ICD-10 codes for underlying causes of death from cancers of the digestive system*

Disaggregating the ICD-10 cancer sections was limited by the small numbers in some sections. We therefore focused on cancers of the digestive system, the most frequently reported cancer-related underlying cause of death in adults with intellectual disabilities and more frequently reported in adults with intellectual disabilities than the general population. Three types of cancer accounted for 82% of cancers of the digestive system in adults with intellectual disabilities (Table 6).

The most frequently recorded was cancer of the colon, rectum and anus – almost half (48%) (n=147) of all cancers of the digestive system were in these sites, a significantly greater proportion than the 34% in the general population ( $X^2(5, N=309) =21.52, p=.00064$ ). The second most frequently recorded cancer of the digestive system in adults with intellectual disabilities was cancer of the oesophagus (19% (n=59) of all cancers of the digestive system), slightly more but not significantly so to the 16% in the general population. Third in adults with intellectual disabilities was cancer of the pancreas (15% (n=45) of all cancers of the digestive system), slightly less, but not significantly so, than the proportion in the general population (20%).

*Potentially avoidable deaths from cancer*

For those in Group 3 who died with cancer as an underlying cause of death, we examined the proportion of cancers known to be avoidable (either preventable or treatable), using the harmonised definition of avoidable mortality and a list of causes of death considered to be avoidable.[21] Comparative information for the general population of England was not available.

Overall, 19% (n=163) adults with intellectual disabilities had a type of cancer that was a preventable cause of death. The largest proportion was due to cancer of the oesophagus (n=52; 32% of all preventable causes of death from cancer) and the lung (n=44; 27% of all preventable causes of death from cancer). 23% (n=200) of adults with intellectual disabilities had a type of cancer that was a treatable underlying cause of death. The largest proportion was due to colorectal cancer (n=106; 53% of all treatable causes of death from cancer) and breast cancer (in females only) (n=57; 29% of all treatable causes of death from cancer).

**Table 6: The most frequently recorded ICD-10 codes for underlying cause of death of cancer of the digestive system for adults with intellectual disabilities and general population of England**

		LeDeR data (age 18+)						England (2018) (age 20+) <sup>‡</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
ICD-10 codes	Neoplasm of...	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers
C18-C21	Colon, rectum and anus	86	45%	60	51%	147	48%	7,509	32%	6,387	37%	13,896	34%
C15	Oesophagus	43	23%	16	14%	59	19%	4,564	20%	1,985	11%	6,549	16%
C25	Pancreas	27	14%	18	15%	45	15%	4,163	18%	3,868	22%	8,031	20%
C22	Liver and intrahepatic bile ducts	15	8%	§	§	23	7%	2,907	12%	1,794	11%	4,701	12%
C16	Stomach	12	6%	§	§	17	6%	2,197	9%	1,213	7%	3,410	8%
C17 C23 C24 C26	All other digestive organs	§	§	10	4%	18	6%	1,957	8%	2,031	12%	3,988	10%
C15-C26	Digestive organs	191	100%	117	100%	309	100%	23,297	100%	17,278	100%	40,575	100%

<sup>†</sup>There is one person whose gender was not recorded. They have been included in the Total column.  
<sup>‡</sup> Data extracted from <https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018>  
<sup>§</sup>Numbers fewer than 10 have been suppressed.

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**DISCUSSION**

Despite the first major study of cancer in people with intellectual disabilities in England being conducted in 1997, the lack of contemporary data leads to significant policy and practice gaps. This study aimed to extend our knowledge about cancer in deceased adults with intellectual disabilities in England. One of the strengths of our study is that it links data about adults with intellectual disabilities known to have had cancer prior to their death with data from the national cancer registry and official cause of death coding from NHS Digital. This provides more comprehensive information than would otherwise be available from any of the data sources alone. There is an indication, however, that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an ‘unknown’ tumour type. The study population was unique in that all died between 2017-2019; it was not a cross-sectional cohort of the population, so comparison with other findings need to be made with this caveat in mind. There was limited published data available about decedents with cancer, so comparing our data about routes to diagnosis and stage of cancer with general population data was not possible. The small number of deaths in some sub-categories, and when compared with general population data about underlying causes of death means that conclusions based on these data should be considered tentative.

Overall, deceased adults with intellectual disabilities had more frequently had cancer diagnosed via emergency presentations than any other routes, suggesting that symptoms suggestive of cancer are not always picked up and acted upon in general practice for adults with intellectual disabilities. Almost half (46%) of cancers in adults with intellectual disabilities were diagnosed at Stage IV when the cancer had already metastasised to other parts of the body. Cancer survival data emphasise the importance of early diagnosis and that for most cancers, survival at one and five years is much higher if the cancer is detected at Stage I than if it is detected later.[24] The NHS Long Term Plan[25] states the ambition that by 2028, the proportion of cancers diagnosed at Stages I and II will rise to three-quarters of cancer patients. With only a third (34%) of cancers in adults with intellectual disabilities identified at these stages, there is clearly much work to do to raise greater awareness of symptoms of cancer in this population, lower the threshold for referral by GPs, and accelerate access to diagnosis and treatment.

Digestive system cancers were the most common type of cancer death in adults with intellectual disabilities in this study, echoing the findings of other studies.[6,8,26] This may be influenced by gastrointestinal tract dysfunction, gastro-oesophageal reflux disease, and chronic constipation which are common in people with intellectual disabilities.[27] There is also a suggestion that a higher incidence of these cancers in adults with intellectual disabilities, compared to the general population, may be associated with genetic deletions and family history, being overweight, inactive, or having poor nutrition.[28] A greater awareness of this amongst support staff and health professionals is important so that preventative measures can be instigated such as caregivers being vigilant about bowel

habits and implementing interventions to increase fibre intake and exercise in daily activities.[28]

Colorectal screening presents an opportunity to discover early colorectal cancer and is available to everyone over the age of 60 years in England, with the programme expanding to include 56-year-olds from 2021. Although screening rates for colorectal cancer in adults with intellectual disabilities are approaching those of the general population[29] our study found that 43% of adults diagnosed with colorectal cancer were aged 18-59 years, suggesting that the age threshold for colorectal screening in people with intellectual disabilities may need reducing.

After colorectal cancer, breast cancer was the second most frequently reported treatable cause of death in our population. Although rates of breast cancer appear to be similar in women with and without intellectual disabilities, under-utilisation of breast cancer screening in adults with intellectual disabilities[29] may reduce the number identified. Nulliparity, being overweight, and a lack of exercise are known factors that increase breast cancer risk[30] and are particularly pertinent to people with intellectual disabilities. Research is sparse about the treatment options and decision-making process for women with intellectual disabilities who have breast cancer; research relating to disabled women in general suggests that they are less likely to undergo breast-conserving surgery and are less likely to receive neoadjuvant chemotherapy and radiotherapy than their non-disabled peers.[31]

Some of the factors influencing disparities in cancer outcomes will be related to the social and economic context that shapes a person's ability to access cancer care. Mechanisms leading to poorer outcomes in people with intellectual disabilities have been identified as provider bias and ableism;[32] negative attitudes;[33] the invisibility of people with intellectual disabilities;[34] diagnostic overshadowing in which symptoms are misinterpreted as due to a person's behaviour, mental state or communication impairments;[35] and unequal access to health services.[36] Preventative measures to reduce the risk of cancer, screening and health checks to identify it early, transparent decision-making processes about options available, and access to diagnostic and treatment interventions that provide a chance of optimal outcomes for people with cancer are all needed. So too, is a closer research gaze on the quality of care provided to people with intellectual disabilities who have cancer in order to ensure their equitable access to services.

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**CONTRIBUTORSHIP STATEMENT**

PH conceived the idea for the study. AC, RC, JP and BS conducted the statistical analyses. PH, was responsible for the initial draft of the report. PH, AC, RC, JP, BS and VB contributed to all subsequent and the final draft. PH and AC act as guarantors for the final manuscript.

**COMPETING INTERESTS**

The authors have declared no competing interests.

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**DATA SHARING STATEMENT**

No data are available.

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STROBE Statement—Checklist of items that should be included in reports of *cross-sectional studies*

	Item No	Recommendation	Page No
Title and abstract	1	(a) Indicate the study’s design with a commonly used term in the title or the abstract	Abstract Line 4 (p.2)
		(b) Provide in the abstract an informative and balanced summary of what was done and what was found	Abstract
Introduction			
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported	p.2-3
Objectives	3	State specific objectives, including any prespecified hypotheses	p.3
Methods			
Study design	4	Present key elements of study design early in the paper	p.3-4
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection	p.3-4
Participants	6	(a) Give the eligibility criteria, and the sources and methods of selection of participants	p.3-4
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable	p.3-4
Data sources/ measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group	p.5
Bias	9	Describe any efforts to address potential sources of bias	p.15
Study size	10	Explain how the study size was arrived at	p.4-5
Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why	p.3
Statistical methods	12	(a) Describe all statistical methods, including those used to control for confounding	p.5
		(b) Describe any methods used to examine subgroups and interactions	p.5
		(c) Explain how missing data were addressed	p.5
		(d) If applicable, describe analytical methods taking account of sampling strategy	n/a
		(e) Describe any sensitivity analyses	n/a
Results			
Participants	13*	(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed	p.6-15
		(b) Give reasons for non-participation at each stage	n/a
		(c) Consider use of a flow diagram	n/a
Descriptive data	14*	(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders	p.6-7
		(b) Indicate number of participants with missing data for each variable of interest	p.6-15

Outcome data	15*	Report numbers of outcome events or summary measures	p.6-15
Main results	16	(a) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included	p.6-15
		(b) Report category boundaries when continuous variables were categorized	n/a
		(c) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period	n/a
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses	p.6-15
<b>Discussion</b>			
Key results	18	Summarise key results with reference to study objectives	p.13-15
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias	p.15-17
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence	p.15-17
Generalisability	21	Discuss the generalisability (external validity) of the study results	p.15-17
<b>Other information</b>			
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based	p.17

\*Give information separately for exposed and unexposed groups.

**Note:** An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at <http://www.plosmedicine.org/>, Annals of Internal Medicine at <http://www.annals.org/>, and Epidemiology at <http://www.epidem.com/>). Information on the STROBE Initiative is available at [www.strobe-statement.org](http://www.strobe-statement.org).

# BMJ Open

## Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

Journal:	<i>BMJ Open</i>
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<b>Primary Subject Heading</b>:	General practice / Family practice
Secondary Subject Heading:	Medical management
Keywords:	Adult oncology < ONCOLOGY, Health policy < HEALTH SERVICES ADMINISTRATION & MANAGEMENT, PRIMARY CARE

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# Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

## TITLE PAGE

### Title

Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

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**ABSTRACT**

**Objective.** To improve our understanding of cancer in adults with intellectual disabilities.

**Design.** Population-based study using linked data about deceased adults from the Learning [Intellectual] Disabilities Mortality Review (LeDeR) programme, the national cancer registry, and NHS Digital.

**Setting.** England.

**Participants.** 1,096 adults with intellectual disabilities identified by the LeDeR programme who died between 1st January 2017 and 31st December 2019.

**Outcome measure.** Any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Parts I or II of the Medical Certificate of Cause of Death.

**Results.** In decedents with intellectual disabilities and cancer, more than a third (35%; n=162) had cancer diagnosed via emergency presentations. Almost half (45%; n=228) of cancers were at Stage IV when diagnosed. More than a third (36%; n=309) of underlying causes of deaths were of cancers of the digestive system; almost half of these (48%; n=147) were cancer of the colon, rectum or anus. Of those who died with colorectal cancer, 43% were below the age threshold for colorectal screening.

**Conclusions.** In decedents with intellectual disabilities, symptoms suggestive of cancer had tended to be identified most frequently as an emergency and at a late stage. There is a need for greater awareness of symptoms of cancer in this population, a lower threshold for referral by GPs, accelerated access to diagnosis and treatment, and consideration paid to lowering the age for colorectal screening.

**STRENGTHS AND LIMITATIONS OF THE STUDY**

- This study is the first to link data about adults with intellectual disabilities known to have had cancer prior to their death with that from the national cancer registry and official cause of death coding from NHS Digital.
- There is an indication that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an ‘unknown’ tumour type.
- There was limited published data available about decedents with cancer, so comparing our data to general population data was not always possible.
- The small number of deaths in some sub-categories, and when compared with general population data about underlying causes of death means that conclusions based on these data should be considered tentative.

## MAIN TEXT

### INTRODUCTION

We have little contemporary data about the experiences of adults with intellectual disabilities who have been diagnosed with cancer.[1] In general, mortality studies of people with intellectual disabilities indicate a lower proportion of cancer-related deaths than in the general population,[2,3] possibly due to the lower life expectancy of people with intellectual disabilities,[4,5] and cancer being predominantly a disease of older age. Nevertheless, cancer is one of the five most frequently recorded causes of death in studies of adults, or adults and children, with intellectual disabilities,[2,6] and is therefore deserving of attention in any considerations about reducing premature mortality in this population.

Within the population of people with intellectual disabilities, the risk of dying from cancer is not uniform. Females with intellectual disabilities may have increased risk of death from cancer compared to males,[2,7] although this mainly appears to be a feature of younger age groups.[6] There are conflicting findings as to whether overall cancer incidence varies by level of intellectual disability. Landes et al.,[2] reported rates of death from cancer to be higher among adults with mild or moderate intellectual disabilities than in adults with severe or profound and multiple intellectual disability, or those with unspecified intellectual disability. Patja et al.,[8] found there to be no difference in overall cancer incidence by level of intellectual disability.

Conclusions have been drawn about the need to improve preventative measures such as cancer screening [9,10] in people with intellectual disabilities, but there has been little published evidence about the diagnosis, treatment and quality of care provided for people with intellectual disabilities with cancer. In a small study of 11 women with intellectual disabilities who had breast cancer, Satgé et al.,[11] observed that cancers were found at a more advanced stage than in the general population. More recently, a population-based cohort study in the Netherlands concluded that cancer may be underdiagnosed and/or undertreated in people with intellectual disability.[12] A scoping review of cancer treatment and survivorship among people with intellectual and developmental disabilities in the U.S. reported that urgent action was needed to improve collaboration among health care providers.[13]

This paper aims to improve our understanding of cancer in adults with intellectual disabilities who have died and whose deaths were reported to the English Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme. It addresses three research questions:

1. What are the types of cancer diagnosed in adults with intellectual disabilities who have died?
2. How, and at what stage, was cancer diagnosed in adults with intellectual disabilities who have died?

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3. What is the underlying cause of death in adults with intellectual disabilities known to have had cancer?

**METHODS**

**Study design and data**

Data about deceased adults with intellectual disabilities were extracted from the LeDeR programme reviews of deaths. The national Learning from Deaths policy[14] requires that all deaths of people with intellectual disabilities (aged 4 years and older) in England should be reported to the LeDeR programme and reviewed using LeDeR programme methodology. The review process is described by [Anonymised] et al.[15]

Data about the ICD-10 codes for causes of death of people with intellectual disabilities notified to the LeDeR programme were obtained from NHS Digital. Data linkage was conducted by NHS Digital and based on the key identifiers of NHS number, date of birth and date of death.

For adults with intellectual disabilities for whom a LeDeR review of their death had been completed and who were known to have had cancer, data was linked to that held in the national cancer registry in England, which holds information about cancer and tumour diseases. Data linkage was conducted by the National Cancer Registration and Analysis Service (NCRAS) and based on the key identifiers of NHS number and date of birth. Those included were adults with intellectual disabilities who died in 2017, 2018 or 2019, for whom a LeDeR review had been completed, and for whom the ICD-10 codes for cause of death had been supplied by NHS Digital. All had cancer listed as a long-term health condition by the reviewer of their death or had cancer included in either Part I or Part II of the Medical Certificate of Cause of Death (MCCD).

Data about decedents in the general population of England were drawn from data already published by the Office for National Statistics in the population data series about deaths, and Public Health England cancer mortality data. Comparative data about other aspects covered in this paper were not available for decedents in England.

The 10th edition of the International Classification of Diseases (ICD-10) was used to classify diagnoses and causes of death. This is divided into 21 chapters; Chapter 2 relates to neoplasms (cancer) with the codes C00-D49.

The ‘stages’ of cancer, which describe the size of the cancer and its spread, are as described by Cancer Research UK[16] as follows:

- Stage I – the cancer is small and has not spread.
- Stage II – the cancer has grown but has not spread.
- Stage III – the cancer is larger and may have spread to the surrounding tissues and/or the lymph nodes.



- Stage IV – the cancer has spread from where it started to at least one other body organ; known as "secondary" or "metastatic" cancer.

Stage 0 refers to 'carcinoma in situ', 'precancerous changes' or 'non-invasive cancer'. Many of these will never develop into cancer and for this reason we have not included them in this study.

We used the World Health Organisation definition of the underlying cause of death: the disease or injury which initiated the train of events leading directly to death[17]. Although we are aware that the underlying cause of death in some people with intellectual disabilities may be inaccurately reported [18, 19, 20], such evidence does not specifically implicate the reporting of deaths from cancer, so we did not amend any cause of death reports.

We used the European harmonised definition of avoidable mortality and list of causes of death that has been adopted by the UK Office for National Statistics:[21]

- Preventable mortality. Causes of death that can be mainly avoided through effective public health and primary prevention interventions (i.e., before the onset of diseases/injuries, to reduce incidence).
- Treatable mortality. Causes of death that can be mainly avoided through timely and effective health care interventions, including secondary prevention and treatment (i.e., after the onset of diseases, to reduce case-fatality).
- Avoidable mortality. Avoidable causes of deaths are all those defined as preventable or treatable.

## Outcome

The outcome of interest was any form of cancer listed as a long-term health condition by a LeDeR reviewer, or ICD-10 codes C00-D49 included on Part I or Part II of the MCCD, of deaths occurring during the period 1<sup>st</sup> January 2017 to 31<sup>st</sup> December 2019.

## Exposure

The definition of intellectual disabilities used was the presence of a significantly reduced ability to understand new or complex information, to learn new skills (impaired intelligence), with a reduced ability to cope independently (impaired social functioning) which started before adulthood, with a lasting effect on development.[22]

## Covariates

The covariates and their sources were:

LeDeR review of death: demographic information - age; gender; ethnicity; level of intellectual disabilities; geographic area of residence; usual type of accommodation.

NHS Digital: ICD-10 codes for causes of death recorded on the MCCD.

National cancer registry: relevant timescales (e.g., patient age at diagnosis); how the cancer was diagnosed; the site and stage of the cancer when it was diagnosed.

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**Ethical approval for the study**

The LeDeR programme had Section 251 approval from the national Health Research Authority’s Confidentiality Advisory Group (CAG), on behalf of the Secretary of State, allowing the programme team to handle identifiable data without consent in order to conduct a review of a death, and to link it to NHS Digital cause of death data. An amendment to the agreement (CAG Ref: 20CAG067) enabled the LeDeR programme to link identifiers with national cancer registry data.

The study was funded by NHS England. NHS England had no part in the analysis or presentation of the data.

**Patient and public involvement**

Family members of people with intellectual disabilities were involved in the conceptualisation of the study and in discussing the findings.

**Data analysis**

Data analysis was undertaken by analysts at [Anonymised]. The analysts worked with the LeDeR team at [Anonymised] to ensure a full understanding of the data and to agree how it was reported. Statistical analyses were carried out using the R programming language. Initial chi-square analyses were carried out on frequency tables as a whole. If there was a significant effect, pairwise chi-square analyses were performed to determine which particular variables had significantly different proportions. For brevity we present only significant pair-wise analyses results.

All numbers fewer than 10 have been suppressed to protect confidentiality.

Where appropriate, data about underlying causes of death have been compared with published general population data.

We present the findings for three distinct but inter-related groups of adults with intellectual disabilities:

Group 1: Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1,096).

Group 2: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771).

Group 3: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852).

**RESULTS**

Demographic data about adults with intellectual disabilities included in each group of the study are presented in Table 1. Overall, there was little difference in the demographic information relating to those in Groups 1, 2 and 3, although the median age at death of those in Group 3 was slightly lower than that of Group 1 and Group 2.

**Table 1: Demographic information about those included in the study**

<b>Group 1:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1,096).						
<b>Group 2:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771).						
<b>Group 3:</b> Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852).						
	<b>Group 1</b>		<b>Group 2</b>		<b>Group 3</b>	
	<b>No.</b>	<b>%</b>	<b>No.</b>	<b>%</b>	<b>No.</b>	<b>%</b>
<b>Gender†</b>						
Males	581	53%	416	54%	457	54%
Females	514	47%	354	46%	394	46%
<b>Age Group</b>						
20-34	44	4%	31	4%	38	4%
35-49	123	11%	75	10%	104	12%
50-64	430	39%	322	42%	369	43%
65-79	420	38%	296	38%	300	35%
80+	79	7%	47	6%	41	5%
<i>Total</i>	<i>1096</i>	<i>100%</i>	<i>771</i>	<i>100%</i>	<i>852</i>	<i>100%</i>
<b>Median age of death</b>	63		63		61	
<b>Standard Deviation</b>	13.33		13.34		12.78	
<b>Ethnicity</b>						
White British	1034	96%	728	96%	803	96%
Non-white British	45	4%	32	4%	35	4%
<i>Total</i>	<i>1079</i>	<i>100%</i>	<i>760</i>	<i>100%</i>	<i>838</i>	<i>100%</i>
<i>Unknown/Missing</i>	<i>17</i>		<i>11</i>		<i>14</i>	
<b>Level of Intellectual Disability</b>						
Mild	453	44%	339	47%	360	45%
Moderate	345	34%	238	33%	268	34%
Severe	183	18%	112	16%	140	18%
Profound / multiple	44	4%	26	4%	31	4%
<i>Total</i>	<i>1025</i>	<i>100%</i>	<i>715</i>	<i>100%</i>	<i>799</i>	<i>100%</i>
<i>Unknown/Missing</i>	<i>71</i>		<i>56</i>		<i>53</i>	
<b>Geographic Area</b>						
Midlands	214	20%	147	19%	171	20%
North East and Yorkshire	168	15%	123	16%	131	15%
South East	181	17%	123	16%	147	17%
North West	160	15%	111	14%	125	15%
East of England	139	13%	92	12%	107	13%
London	128	12%	96	12%	94	11%
South West	106	10%	79	10%	77	9%
<i>Total</i>	<i>1096</i>	<i>100%</i>	<i>771</i>	<i>100%</i>	<i>852</i>	<i>100%</i>
<b>Accommodation</b>						
Supported living	258	33%	163	32%	198	32%
Own or Family	207	27%	137	27%	175	28%
Residential home	190	24%	122	24%	145	23%
Nursing home	109	14%	67	13%	89	14%
Other	14	2%	11	2%	11	2%
<i>Total</i>	<i>778</i>	<i>100%</i>	<i>500</i>	<i>100%</i>	<i>618</i>	<i>100%</i>

<i>Unknown/Missing</i>	318	271	234
†There is one person whose gender was not recorded. ‡Numbers fewer than 10 have been suppressed.			

**Adults with intellectual disabilities known from LeDeR data to have died with cancer (Group 1)**

There were 1,096 adults with intellectual disabilities who were known from LeDeR data to have died with cancer. Demographic data for adults with intellectual disabilities in Group 1 were similar to the overall adult population of people with intellectual disabilities whose deaths were reported to the LeDeR programme in 2018.[23]

Most (70%; n=771) of those included in Group 1 had linked data available from the national cancer registry and from Group 2. However, 325 (30%) adults were known by the LeDeR programme to have died with cancer, but data was not available about them in the national cancer registry.

The majority of those for whom data in the national cancer registry were not available (70%, n=229) had died in 2019, a year for which registrations in the national cancer registry had not yet been completed at the time of writing. Thus, it is likely that these deaths were late registrations rather than omissions from the registry.

Data were not available from the national cancer registry for 96 (9%) adults who died in 2017 or 2018 and who were known by the LeDeR programme to have had cancer. These people tended to be older (16% aged 80 or over, compared to 6% of those in the registry); and more had severe or profound and multiple intellectual disabilities (36%) than those in the registry (19%). A quarter (25%) of those not in the cancer registry had an ‘unknown’ tumour type, compared to none of those included.

**Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2)**

There were 771 adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2).

Information about the type of cancer diagnosed was available for all 771 of those in Group 2 (Table 2). Most (89%, n=690) had been diagnosed with one type of cancer; 9% (n=69) with two; 2% (n=12) with three or more different types. Thus, in the 771 people, 865 cancers had been diagnosed.

In males, the most frequently recorded types of cancers were of the digestive organs (28%); skin (12%); lip, oral cavity and pharynx (11%); and male genital organs (10%). In females, the most frequently recorded types of cancer were of the breast (26%); digestive organs (23%); and female genital organs (14%).

**Table 2: The most frequently reported cancers by ICD-10-chapter sections in adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2) (n=771)**

ICD-10 section	Section heading	Males		Females		Total <sup>†</sup>	
		No	%	No	%	No	%
C15-C26	Malignant neoplasms of digestive organs	131	28%	92	23%	224	26%
C50	Malignant neoplasms of breast	§	§	103	26%	105	12%
C43-C44	Melanoma and other malignant neoplasms of skin	56	12%	32	8%	88	10%
C00-C14	Malignant neoplasms of lip, oral cavity and pharynx	52	11%	13	3%	65	8%
C30-C39	Malignant neoplasms of respiratory and intrathoracic organs	39	8%	21	5%	60	7%
C42	Malignant neoplasms of the haemato-poietic and reticuloendothelial system	33	7%	22	6%	55	6%
C51-C58	Malignant neoplasms of female genital organs			54	14%	54	6%
C60-C63	Malignant neoplasms of male genital organs	47	10%			47	5%
C64-C68	Malignant neoplasms of urinary tract	33	7%	13	3%	46	5%
C76-C80	Malignant neoplasms of ill-defined, other secondary and unspecified sites	24	5%	13	3%	37	4%
C81-C96	Malignant neoplasms of lymphoid, hematopoietic and related tissue	19	4%	15	4%	34	4%
C69-C72	Malignant neoplasms of eye, brain and other parts of central nervous system	20	4%	10	3%	30	3%
C40-41; C45-49; C73-75; D00-09; D10-36 D37-48; D49	All other cancers.	11	2%	§	§	20	2%
<b>Total number of cancers in the 771 people</b>		<b>467</b>	<b>100%</b>	<b>397</b>	<b>100%</b>	<b>865</b>	<b>100%</b>

<sup>†</sup>There is one person whose gender was not recorded. They have been included in the Total column.  
<sup>§</sup>Numbers fewer than 10 have been suppressed.

Of the 771 adults with intellectual disabilities in Group 2, information about the route to diagnosis was available for 60% (n=462). In these 462 adults with intellectual disabilities, cancers were more likely to be diagnosed via emergency presentations than any other route: 35% of adults with intellectual disabilities had their cancer diagnosed via an

emergency referral or attendance, compared to 27% diagnosed via a non-urgent referral by their GP, and 25% diagnosed via an urgent referral (Table 3).

Table 3: Route to diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry		
	LeDeR data (2017-2019)	
	No.	%
Emergency presentation	162	35%
GP non-urgent referral	123	27%
Urgent referral ('two-week wait')	116	25%
Other (Outpatient attendance/ elective inpatient/ death certificate only)	45	10%
Screening	16	3%
Total	462	100%
Unknown/Missing	309	

Adults with intellectual disabilities who were diagnosed via an emergency route were disproportionately male (20%, compared to 15% females;  $\chi^2(5, N=462) = 29.95, p=.0042$ ); younger in age (51% in the 20-49 age group, compared to 33% of those in older age groups;  $\chi^2(20, N=462) = 26.65, p=.0017$ ); and living in their own or the family home (36%, compared to 16% of those living in residential care setting;  $\chi^2(25, N=462) = 64.68, p=.0014$ ).

Sixteen adults with intellectual disabilities had their cancer identified by screening. Of these, 14 were diagnosed with breast cancer, a rate comparable with the proportion of cancers identified by screening in the general population.

Of the 865 cancers diagnosed in the 771 adults with intellectual disabilities in Group 2, information about the stage of the cancer when it was diagnosed was available for 58% (n=502) of the different cancers. Almost half (46%) of the cancers diagnosed were at Stage IV when the cancer had already metastasised; two thirds (66%) were at Stage III or IV (Table 4).

Table 4: Stage of cancer at diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry		
	LeDeR data (2017-2019)	
	No.	%
Stage I	88	18%
Stage II	78	16%
Stage III	105	21%



Stage IV	228	45%
Total number of cancers	502	100%
Unknown/Missing/unstageable	363	

There was some variation in the stage of cancer in adults with intellectual disabilities in Group 2. Those cancers diagnosed at Stage IV were disproportionately in males (51%, compared to 39% in females;  $\chi^2 (8, N=503) = 15.92, p=.0033$ ); in adults in younger age groups (52% aged 20-49 years, compared to 45% in older age groups  $\chi^2 (16, N=503) = 27.18, p=.002$ ); and of cancer of the digestive organs (57% of cancers of the digestive system were diagnosed at Stage IV;  $\chi^2 (56, N=503) = 265.9, p=.0007$ ).

### **Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (Group 3)**

#### *ICD-10 chapters of underlying causes of death*

Over three-quarters ( $n=852$ ; 78%) of the 1,096 adults with intellectual disabilities in Group 1 had cancer recorded as their underlying cause of death in the official cause of death coding received from NHS Digital. Small proportions had respiratory disorders (7%;  $n=73$ ), circulatory disorders (4%;  $n=42$ ) or other disorders (11%;  $n=124$ ) recorded as their underlying cause of death.

#### *ICD-10 sections of cancer-related underlying causes of death*

The most frequently recorded ICD-10 section for the cancer-related underlying cause of deaths in the 852 adults in Group 3 was cancer of the digestive organs (C15-26), reported for over a third (36%) of deaths (Table 5). The corresponding proportion in decedents in the general population of England was 29%. The second most frequently recorded in adults with intellectual disabilities in Group 3 was of ill-defined, secondary and unspecified sites (10%), more than the corresponding proportion in the general population of England (6%), and possibly reflecting the greater than expected proportion of cancers in adults with intellectual disabilities that were diagnosed at emergency presentation and in the later stages. The third most frequently recorded in adults with intellectual disabilities in Group 3 was of cancer of the respiratory and intrathoracic organs (8%), although this was substantially less than in the general population of England (21%).

There was some variation within the adults with intellectual disabilities in Group 3 according to the underlying cause of death. Cancers of the digestive system were more frequently reported in males than females (42% males; 30% females;  $\chi^2 (34, N=852) = 209.63, p=.0002$ ); and cancers of the male genital organs were not reported in any males from minority ethnic groups ( $\chi^2 (34, N=852) = 60.262, p=.0011$ ).

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We compared the most frequently recorded ICD-10 sections for the cancer-related underlying causes of death for adults with intellectual disabilities in Group 3 with those of the general population of England using a chi-square test. There was a significant difference

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**Table 5: The most frequently recorded ICD-10 codes for cancer-related underlying causes of death in adults with intellectual disabilities for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (Group 3), compared to data reported for the general population of England**

ICD-10 codes	Neoplasm of...	LeDeR data (2017-2019) (age 18+)						General population (2018) (age 20+) <sup>±</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
		no.	%	no.	%	no.	%	no.	%	no.	%	no.	%
C15-C26	Digestive organs	191	42%	117	30%	309	36%	23,297	32%	15,953	26%	39,250	29%
C76-C80	Ill-defined, other secondary & unspecified sites	53	12%	32	8%	85	10%	4,003	6%	4,448	7%	8,451	6%
C30-C39	Respiratory & intrathoracic organs	46	10%	23	6%	69	8%	15,708	22%	12,910	21%	28,618	21%
C50	Breast	§	§	67	17%	69	8%	78	<1%	9,562	15%	9,640	7%
C81-C96; C42	Lymphoid, hematopoietic & related tissue	37	8%	27	7%	64	8%	3,799	5%	3,233	5%	7,032	5%
C51-C58	Female genital organs			60	15%	60	7%			6,818	11%	6,818	5%
C64-C68	Urinary tract	32	7%	18	5%	50	6%	5,492	8%	2,844	5%	8,336	6%
C69-C72	Eye, brain & other central nervous system	24	5%	12	3%	36	4%	2,200	3%	1,603	3%	3,803	3%
C60-C63	Male genital organs	34	7%			34	4%	10,243	14%			10,243	8%
C43-C44	Melanoma & other skin	17	4%	13	3%	30	4%	1671	2%	1,007	2%	2,678	2%
C00-14; C40-41;	All other causes	22	5%	24	6%	46	5%	5,935	8%	3,691	5%	9,626	8%

C45-49; C73-75; D00-09; D10-36; D37-48.													
<b>Total</b>		458	100%	393	100%	852	100%	72,426	100%	62,069	100%	134,495	100%
<p>†There is one person whose gender was not recorded. They have been included in the Total column.</p> <p>‡ Data extracted from <a href="https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018">https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018</a></p> <p>§Numbers fewer than 10 have been suppressed.</p>													

between the population of adults with intellectual disabilities and adults in the general population ( $X^2(10, N=852) = 21.79, p=.016$ ). Pairwise testing suggested that in males with intellectual disabilities, genital cancer and respiratory cancer were significantly lower than in males in the general population. In females with intellectual disabilities, respiratory cancer was significantly lower than in females in the general population.

### *Specific ICD-10 codes for underlying causes of death from cancers of the digestive system*

Disaggregating the ICD-10 cancer sections was limited by the small numbers in some sections. We therefore focused on cancers of the digestive system, the most frequently reported cancer-related underlying cause of death in adults with intellectual disabilities and more frequently reported in adults with intellectual disabilities than the general population. Three types of cancer accounted for 82% of cancers of the digestive system in adults with intellectual disabilities (Table 6).

The most frequently recorded was cancer of the colon, rectum and anus – almost half (48%) (n=147) of all cancers of the digestive system were in these sites, a significantly greater proportion than the 34% in the general population ( $X^2(5, N=309) = 21.52, p=.00064$ ). The second most frequently recorded cancer of the digestive system in adults with intellectual disabilities was cancer of the oesophagus (19% (n=59) of all cancers of the digestive system), slightly more but not significantly so to the 16% in the general population. Third in adults with intellectual disabilities was cancer of the pancreas (15% (n=45) of all cancers of the digestive system), slightly less, but not significantly so, than the proportion in the general population (20%).

### *Potentially avoidable deaths from cancer*

For those in Group 3 who died with cancer as an underlying cause of death, we examined the proportion of cancers known to be avoidable (either preventable or treatable), using the harmonised definition of avoidable mortality and a list of causes of death considered to be avoidable.[21] Comparative information for the general population of England was not available.

Overall, 19% (n=163) adults with intellectual disabilities had a type of cancer that was a preventable cause of death. The largest proportion was due to cancer of the oesophagus (n=52; 32% of all preventable causes of death from cancer) and the lung (n=44; 27% of all preventable causes of death from cancer). 23% (n=200) of adults with intellectual disabilities had a type of cancer that was a treatable underlying cause of death. The largest proportion was due to colorectal cancer (n=106; 53% of all treatable causes of death from cancer) and breast cancer (in females only) (n=57; 29% of all treatable causes of death from cancer).

Table 6: The most frequently recorded ICD-10 codes for underlying cause of death of cancer of the digestive system for adults with intellectual disabilities and general population of England													
		LeDeR data (age 18+)						England (2018) (age 20+) <sup>‡</sup>					
		Male		Female		Total <sup>†</sup>		Male		Female		Total	
ICD-10 codes	Neoplasm of...	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers	no.	% of digestive cancers
C18-C21	Colon, rectum and anus	86	45%	60	51%	147	48%	7,509	32%	6,387	37%	13,896	34%
C15	Oesophagus	43	23%	16	14%	59	19%	4,564	20%	1,985	11%	6,549	16%
C25	Pancreas	27	14%	18	15%	45	15%	4,163	18%	3,868	22%	8,031	20%
C22	Liver and intrahepatic bile ducts	15	8%	§	§	23	7%	2,907	12%	1,794	11%	4,701	12%
C16	Stomach	12	6%	§	§	17	6%	2,197	9%	1,213	7%	3,410	8%
C17 C23 C24 C26	All other digestive organs	§	§	10	4%	18	6%	1,957	8%	2,031	12%	3,988	10%
C15-C26	Digestive organs	191	100%	117	100%	309	100%	23,297	100%	17,278	100%	40,575	100%
<sup>†</sup> There is one person whose gender was not recorded. They have been included in the Total column.													
<sup>‡</sup> Data extracted from <a href="https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018">https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018</a>													
§Numbers fewer than 10 have been suppressed.													

## DISCUSSION

Despite the first major study of cancer in people with intellectual disabilities in England being conducted in 1997, the lack of contemporary data leads to significant policy and practice gaps. This study aimed to extend our knowledge about cancer in deceased adults with intellectual disabilities in England. One of the strengths of our study is that it links data about adults with intellectual disabilities known to have had cancer prior to their death with data from the national cancer registry and official cause of death coding from NHS Digital. This provides more comprehensive information than would otherwise be available from any of the data sources alone. There is an indication, however, that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an 'unknown' tumour type. We did not check the validity of the MCCD in the study population, but evidence suggests that inaccurate reporting of cause of death of people with intellectual disabilities is less likely when cancer is included as a cause of death.[24] The study population was unique in that all died between 2017-2019; it was not a cross-sectional cohort of the population, so comparison with other findings need to be made with this caveat in mind. There was limited published data available about decedents with cancer, so comparing our data about routes to diagnosis and stage of cancer with general population data was not possible. The small number of deaths in some sub-categories, and when compared with general population data about underlying causes of death means that conclusions based on these data should be considered tentative.

Overall, deceased adults with intellectual disabilities had more frequently had cancer diagnosed via emergency presentations than any other routes, suggesting that symptoms suggestive of cancer are not always picked up and acted upon in general practice for adults with intellectual disabilities. Almost half (46%) of cancers in adults with intellectual disabilities were diagnosed at Stage IV when the cancer had already metastasised to other parts of the body. Cancer survival data emphasise the importance of early diagnosis and that for most cancers, survival at one and five years is much higher if the cancer is detected at Stage I than if it is detected later.[25] The NHS Long Term Plan[26] states the ambition that by 2028, the proportion of cancers diagnosed at Stages I and II will rise to three-quarters of cancer patients. With only a third (34%) of cancers in adults with intellectual disabilities identified at these stages, there is clearly much work to do to raise greater awareness of symptoms of cancer in this population, lower the threshold for referral by GPs, and accelerate access to diagnosis and treatment.

Digestive system cancers were the most common type of cancer death in adults with intellectual disabilities in this study, echoing the findings of other studies.[6,8,27] This may be influenced by gastrointestinal tract dysfunction, gastro-oesophageal reflux disease, and chronic constipation which are common in people with intellectual disabilities.[28] There is also a suggestion that a higher incidence of these cancers in adults with intellectual disabilities, compared to the general population, may be associated with genetic deletions and family history, being overweight, inactive, or having poor nutrition.[29] A greater

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awareness of this amongst support staff and health professionals is important so that preventative measures can be instigated such as caregivers being vigilant about bowel habits and implementing interventions to increase fibre intake and exercise in daily activities.[29]

Colorectal screening presents an opportunity to discover early colorectal cancer and is available to everyone over the age of 60 years in England, with the programme expanding to include 56-year-olds from 2021. Although screening rates for colorectal cancer in adults with intellectual disabilities are approaching those of the general population[30] our study found that 43% of adults diagnosed with colorectal cancer were aged 18-59 years, suggesting that the age threshold for colorectal screening in people with intellectual disabilities may need reducing.

After colorectal cancer, breast cancer was the second most frequently reported treatable cause of death in our population. Although rates of breast cancer appear to be similar in women with and without intellectual disabilities, under-utilisation of breast cancer screening in adults with intellectual disabilities[30] may reduce the number identified. Nulliparity, being overweight, and a lack of exercise are known factors that increase breast cancer risk[31] and are particularly pertinent to people with intellectual disabilities. Research is sparse about the treatment options and decision-making process for women with intellectual disabilities who have breast cancer; research relating to disabled women in general suggests that they are less likely to undergo breast-conserving surgery and are less likely to receive neoadjuvant chemotherapy and radiotherapy than their non-disabled peers.[32]

Some of the factors influencing disparities in cancer outcomes will be related to the social and economic context that shapes a person’s ability to access cancer care. Mechanisms leading to poorer outcomes in people with intellectual disabilities have been identified as provider bias and ableism;[33] negative attitudes;[34] the invisibility of people with intellectual disabilities;[35] diagnostic overshadowing in which symptoms are misinterpreted as due to a person’s behaviour, mental state or communication impairments;[36] and unequal access to health services.[37] Preventative measures to reduce the risk of cancer, screening and health checks to identify it early, transparent decision-making processes about options available, and access to diagnostic and treatment interventions that provide a chance of optimal outcomes for people with cancer are all needed. So too, is a closer research gaze on the quality of care provided to people with intellectual disabilities who have cancer in order to ensure their equitable access to services.

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## CONTRIBUTORSHIP STATEMENT

PH conceived the idea for the study. AC, RC, JP and BS conducted the statistical analyses. PH, was responsible for the initial draft of the report. PH, AC, RC, JP, BS and VB contributed to all subsequent and the final draft. PH and AC act as guarantors for the final manuscript.

## COMPETING INTERESTS

The authors have declared no competing interests.

## FUNDING

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## DATA SHARING STATEMENT

No data are available.

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STROBE Statement—Checklist of items that should be included in reports of *cross-sectional studies*

	Item No	Recommendation	Page No
Title and abstract	1	(a) Indicate the study’s design with a commonly used term in the title or the abstract	Abstract Line 4 (p.2)
		(b) Provide in the abstract an informative and balanced summary of what was done and what was found	Abstract
Introduction			
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported	p.2-3
Objectives	3	State specific objectives, including any prespecified hypotheses	p.3
Methods			
Study design	4	Present key elements of study design early in the paper	p.3-4
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection	p.3-4
Participants	6	(a) Give the eligibility criteria, and the sources and methods of selection of participants	p.3-4
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable	p.3-4
Data sources/measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group	p.5
Bias	9	Describe any efforts to address potential sources of bias	p.15
Study size	10	Explain how the study size was arrived at	p.4-5
Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why	p.3
Statistical methods	12	(a) Describe all statistical methods, including those used to control for confounding	p.5
		(b) Describe any methods used to examine subgroups and interactions	p.5
		(c) Explain how missing data were addressed	p.5
		(d) If applicable, describe analytical methods taking account of sampling strategy	n/a
		(e) Describe any sensitivity analyses	n/a
Results			
Participants	13*	(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed	p.6-15
		(b) Give reasons for non-participation at each stage	n/a
		(c) Consider use of a flow diagram	n/a
Descriptive data	14*	(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders	p.6-7
		(b) Indicate number of participants with missing data for each variable of interest	p.6-15

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Outcome data	15*	Report numbers of outcome events or summary measures	p.6-15
Main results	16	(a) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included	p.6-15
		(b) Report category boundaries when continuous variables were categorized	n/a
		(c) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period	n/a
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses	p.6-15
<b>Discussion</b>			
Key results	18	Summarise key results with reference to study objectives	p.13-15
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias	p.15-17
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence	p.15-17
Generalisability	21	Discuss the generalisability (external validity) of the study results	p.15-17
<b>Other information</b>			
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based	p.17

\*Give information separately for exposed and unexposed groups.

**Note:** An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at <http://www.plosmedicine.org/>, Annals of Internal Medicine at <http://www.annals.org/>, and Epidemiology at <http://www.epidem.com/>). Information on the STROBE Initiative is available at [www.strobe-statement.org](http://www.strobe-statement.org).